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Case Report

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A RARE CASE OF TRIGONOCEPHALY

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ABSTRACT

Premature closure of the metopic suture results in a growth restriction of the frontal bones, which leads to a skull malformation known as trigonocephaly. This report describes our experience of a case of trigonocephaly.

KEYWORD: Trigonocephaly, Metopic Synostosis.

INTRODUCTION

The term trigonocephaly is derived from the Greek words “trigonon”, which means triangle, and “kephale”, which means head. The term trigonocephaly was coined by Welcker in 1862.[1] The metopic suture separates the two frontal bones at birth and is the first skull suture to close physiologically.[2] A premature fusion however, results not only in an obvious ridge over the midline of the forehead, but also in a lateral growth restriction of the frontal bones. According to the theory of Virchow, this wedge shape is even further enhanced by the increased compensatory growth of the remaining skull sutures while the skull keeps expanding.[3] The end product is a skull with a triangular forehead, a bony midline ridge and a shortening of the anterior cranial fossa. In this report we describe a case of a 4 year old who presented with abnormal shape of head and was diagnosed to have trigonocephaly on CT and underwent multiple fontanelle osteotomies with correction of trigonocephaly.

CASE PRESENTATION

A 4 month old female baby born to a non-consanguineous married couple presented with abnormal shape of head since birth. Baby was delivered by lower segment caesarean section. Antenatal period was uneventful. There was no delay in any of the developmental milestones. No history of trauma. On Physical examination Anterior fontanelle was open, frontal bossing was present, posterior fontanelle was closed, parietal prominence was present. CT scan showed features of mild trigonocephaly with bilateral frontal and temporal lobe atrophy, depressed nasal bridge, fused coronal sutures bilaterally. Child was operated for multiple frontal osteotomies with correction of trigonocephaly, with closure of metopic suture and pull up of sphenoid ridge bilaterally. Patient recovered uneventfully postoperatively and was discharged.

Figure 1: Abnormal shape of head.

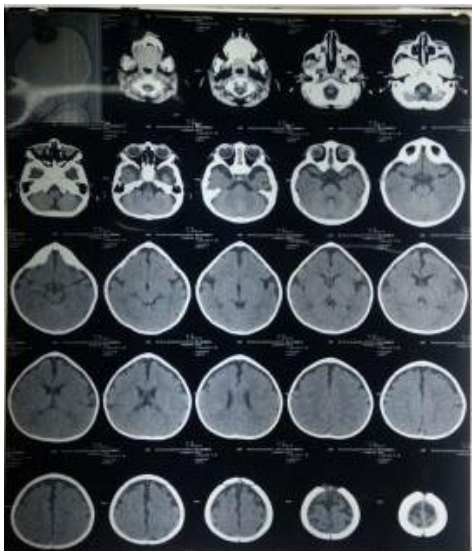

Figure 2: CT scan showing features of mild trigonocephaly.

Figure 3: Post surgery.

DISCUSSION

Trigonocephaly is a congenital cranial anomaly with a narrow pointed forehead, associated with bi parietal widening, which results in a triangular skull with partial or complete obliteration of the metopic sutures. Incidence varies between 1:700- 1:15000 in new born.[4] The male to female ratio is reported to be between 2:1[5] and 6.5:1.[6] They also found a positive family history in 10 out of the 179 families (5.6 %) and a 7.8 % frequency of twins. Increased maternal age and a birth weight of less than 2,500 g was associated with a higher risk of metopic synostosis.[7]

The etiology of metopic synostosis is largely unknown. Following theories are proposed- Intrinsic bone malformation, Fetal head constrain, Intrinsic brain malformation

Observatory signs are: triangular fore head with smaller anterior cranial fossa, visible and palpable midline ridge, hypertelorism including ethmoidal hypoplasia.

Computed tomography (CT) scanning and three dimensional reconstruction using both bone and soft tissue windows is the investigation of choice.[8] MRI and CT scan show epicanthal fold, tear drop- shaped orbits angulated towards midline called surprised coon sign, anterior curving of the metopic sutures, normal cephalic index with bi temporal shortening and bi parietal broadening.

A percentage of patients exhibit neuro psychological signs in the form of behavioural, speech and language problems, mental retardation and neurodevelopmental delays such as attention deficit hyperactive disorder, autism spectrum disorder and conduct deficit.[9]

Treatment is commonly accepted to be surgical. Due to claims of better intellectual outcome, the operative correction is generally performed before the age of one.[10]

CONCLUSION

The optimal time for operative treatment of TRIGONOCEPHALY is generally considered to be the first 6 months after birth Given the high incidence of intracranial abnormalities and neuropathology. Early diagnosis and treatment before the appearance of developmental delay, is proved to be essential.

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